Clinical Records

Unilateral hearing loss due to a rhabdomyoma in a six-year-old child

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Abstract

A case report of a six-year-old child is presented, who had had a unilateral sensorineural hearing loss for several years. Because of impairment in the ABR as well as in the caloric testing a MRI and CT scan were performed. A 17 mm tumour in the cerebello-pontine angle (CPA) was detected, which after suboccipital surgery proved to be a rhabdomyoma. This tumour has not been described before in the CPA. Unilateral sensorineural hearing loss should, at all ages, be an indication for further (radiodiagnostic) investigations.

Key words: Rhabdomyoma; Hearing loss, unilateral; Cerebello-pontine angle; Child

Introduction

Unilateral sensorineural hearing loss and complete unilateral deafness seems to occur in one out of 1000 children of school age (Everberg, 1960). The cause most often remains unknown. Rarely a genetic origin can be traced (Marres, 1994). In adolescents and adults a small asymmetric sensorineural hearing loss requires an investigation of the cerebello-pontine angle to exclude a space-occupying lesion. A case report is presented of a six-year-old boy with a unilateral sensorineural hearing loss. After histological studies it was found to be a rhabdomyoma, which previously has been reported only once in an intracranial location in the trigeminal nerve (Zwick et al., 1989). Nowadays, we have access to advanced imaging techniques of the inner ear and the cerebello-pontine angle, and modern radiology is non-expensive and comfortable for patients these additional investigations should also be considered in childhood.

Case report

In 1990, a six-year-old child was referred to the ORL department of the University Hospital, Nijmegen because of a unilateral hearing loss, found at a regular screening at school. There was no medical history. In a scheduled examination after the first year of life a hearing loss had been noticed, but no further action had been taken. The speech and motor development were normal.

At the ENT-examination a normal eardrum was seen, with air-containing middle ear. The tuning fork test according to Rinne and Weber were normal, and the Barany noise box test proved that both ears could hear. At pure tone audiometry a sensorineural hearing loss was found with a Fletcher Index (mean loss at 0.5, 1.0 and 2.0 kHz) of 50 dB in the left ear. Speech audiometry gave a word discrimination of 100 per cent with the same 50 dB shift. The hearing was normal in the right ear. Contralateral stapedial reflexes could be elicited on both sides. The ABR on the left side revealed an interwave delay from J1-J3 and J1-J5. On the right side a much smaller interwave delay J1-J5 was found. Labyrinth testing with caloric indicates a reduced function of the left labyrinth.

MRI scanning had just become available in a nearby hospital and a tumour of the eighth nerve was found (Figure 1). It was not possible to use GadoLium at that time. Later, a CT scan with the use of contrast showed a tumour of 17 mm in the left cerebello-pontine angle (Figure 2). Angiography showed no abnormal vascular structures.

The patient was referred to the neurosurgical department, and a suboccipital approach was used for the exploration of the cerebello-pontine angle. During the operation the tumour showed encapsulation with a smooth surface and firm consistency. There was an ingrowth in a dilated internal acoustic meatus and adhesions were found with the pons. It was not possible to remove the tumour totally. Post-operatively there was a fast recovery, but a total facial paralysis House grade VI (House and Brackmann, 1985) remained. The left ear was completely deaf. There were no vertigo problems afterwards. MRI-scanning three years after the operation showed no growth of the remaining part of the tumour. Four years after the operation, a cross face nerve transplant was made using the left sural nerve to reinnervate the left facial nerve.

Pathology

During the operation a frozen section examination was performed and because of the presence of matured skeletal muscle a diagnosis of teratoma was suggested. In the permanent paraffin sections the tumour showed large well-developed matured skeletal muscle fibres with easily identifiable haematoxylin and eosin stain cross striations, accentuated by PTAH-Mallory stain (Figure 3). The fibres

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MRI without Gadolinium of the rhabdomyoma in the left cerebello-pontine angle.

were arranged in irregular fascicles and separated by various amounts of fibrocollagenous tissue. No cystic structures, vacuolated cells or spider cells were found. There was no mitotic activity. Muscle fibres showed diffuse cytoplasmic immunoreactivity for desmin, muscle-specific actin HHF-35 and myoglobin. No glial tissue was detected and there was no reaction for S100 protein and GFAP. Some tumour cells stained also for vimentin. The diagnosis rhabdomyoma was made.

Discussion

Hearing loss and even deafness of one ear in childhood is a regular finding in daily practice. Usually, the hearing loss is due to middle ear problems and will be solved in time, by using medication or middle ear ventilation tubes. When a sensorineural hearing loss is found there is a serious indication for further investigations. Tumours as in this young child are rare but can exist for several years giving symptoms which should not be overlooked.

Congenital intracranial tumours are rare and seldomly found in the cerebello-pontine angle (Werb et al., 1992). A rhabdomyoma is defined as a benign and usually circumscribed tumour consisting of mature muscle cells (Kleihues et al., 1993). Other benign mesenchymal tumours with intracranial locations such as leiomyoma of fibrous xanthoma have also been reported (Burger and Scheithauer, 1994).

The location of rhabdomyoma can be cardiac or extracardiac. Cardiac rhabdomyomas are rare congenital tumours resulting from early dysembryogenetic disorder of organogenesis. They are generally benign lesions considered as hamartomas which may be the first manifestation of Bourneville's tuberous sclerosis (Mehta, 1993). The extracardiac rhabdomyomas may assume a number of histological patterns. Tumours composed of uniform large polygonal eosinophilic cells with occasionally cross stria-

CT scan with contrast of the rhabdomyoma in the left cerebello-pontine angle.
of a primary intracranial rhabdomyoma which involved the
We were able to find in the literature only one other case
should be considered as a choristoma or a true neoplasm,
therapy (Taratuto
confirmed the benign character of the lesion. Overall
malignant tumour and patients stay alive no longer than
myosarcomas both extracranially and intracranially
incidence of rhabdomyomas is much lower than rhabdo-
findings were consistent with the rhabdomyomatous
chymal cells, vacuolated or globular rhabdomyoblasts and
mitotic activity. Immature neuroblasts, immature mesen-
Rhabdomyosarcomas have been reported to arise in both
the leptomeninges and the brain parenchyma (Korinthen-
berg et al., 1984; Taratutu et al., 1985; Ferracini et al.,
Focal skeletal muscle ectopia in leptomeninges has
been reported predominantly in children with other
developmental anomalies of the central nervous system
and sometimes chromosomal abnormalities which suggests
faulty mesenchymal differentiation secondary to a genetic
error in the tissue regulation. In several cases skeletal
muscle has been found in leptomeninges of the pontine
region or ponto-medullary junction (Johnson and Ludwin,
1984; Fix et al., 1989). Interestingly, the tumour reported
by us was also located in the pontine angle area.

Conclusion
Unilateral sensorineural hearing loss may be an alarming
symptom in childhood. Further investigations using CT
scanning and MRI should be considered to find the cause
of the phenomenon. In childhood rare congenital tumours
such as a rhabdomyoma are sometimes found in these
cases and early exploration increases the chance of
successful radical surgery. The presented child is alive
and well four years after operation, although only an
incomplete resection could be performed. An additional
surgical procedure for the total facial paralysis was
performed.

Acknowledgement
The authors wish to thank Dr P. C. Burgers (Department
of Pathology, Duke University Medical Centre,
Durham USA) for his opinion on the presented case.

References
Agamanolis, D. P., Dasu, S., Krill, C. E. (1986) Tumors of
unusual forms. Otolaryngologic Clinics of North America
19: 659-683.
Burger, P. C., Scheithauer, B. W. (1994) Tumours of the central
Forces Institute of Pathology, Washington, pp 302-308.
rhabdomyoma: an intermediate form of skeletal muscle
tumor in children. Archives of Pathology and Laboratory
117: 43-47.
Tissue Tumors. Mosby, St. Louis, pp 433-447.
and genetic investigations. Acta Oto-Laryngologica (Stock-
with rhabdomyoblastic differentiation: case report. Neuro-
surgery 30: 782-785.
rhabdomyomatosis of the posterior fossa. Archives of
Pathology and Laboratory Medicine 113: 873-873.
Fu, Y., Perzin, K. H. (1976) Nonepithelial tumors of the nasal
cavity, paranasal sinuses and nasopharynx. Cancer 37:
364-376.

Fig. 3
Photomicrograph of the tumour illustrating well developed
skeletal muscle fibres with easily identified cross striation
accentuated by PTAH-Malory stain. The picture is consistent
with rhabdomyoma (× 250).

trigeminal nerve. The authors regarded the lesion as
choristoma and postulated that the tumour arose from
embryologically disrupted and dislocated masticatory
myogenic tissue following innervation of the mandibular
motor division of the trigeminal nerve (Zwick et al., 1989).
The foci of skeletal muscles can be found accidentally in
normal meninges, but their relationship to rhabdomyomas and
rhabdomyosarcomas is not clear (Hoffman and Rorke,
1971; Fix et al., 1989; Burger and Scheithauer, 1994).
Rhabdomyosarcomas may be encountered as a component of
the malignant germ cell tumours of the pineal gland and
rhabdoid tumours (Jakate et al., 1985; Kodet et al., 1989;
Burger and Scheithauer, 1994). Foci of skeletal muscle
are typically found in infancy and childhood. They may
be located in the leptomeninges and the brain parenchyma (Korinthenberg et al., 1984; Taratutu et al., 1985; Ferracini et al., 1992). The question of whether the presented case
should be considered as a choristoma or a true neoplasm
is a matter of debate. In this case the tumour classified as the adult-type
rhabdomyoma showed other distinctive features and no
mitotic activity. Immature neuroblasts, immature mesenchymal cells, vacuolated or globular rhabdomyoblasts and
glial cells were not found. Our immuno-histochemical
findings were consistent with the rhabdomyomatous
phenotype (Helliwell et al., 1988; Kapadia et al., 1993).
The four year follow-up time free of recurrent disease
confirmed the benign character of the lesion. Overall
incidence of rhabdomyomas is much lower than rhabdo-
myosarcomas both extracranially and intracranially
(Enzinger and Weiss, 1988; Zwick et al., 1989; Burger and Scheithauer, 1994). Rhabdomyosarcoma is a highly
malignant tumour and patients stay alive no longer than
two years despite aggressive chemotherapy and radio-
therapy (Taratuto et al., 1985).
It is a question of debate whether the presented case
should be considered as a choristoma or a true neoplasm.
We were able to find in the literature only one other case
of a primary intracranial rhabdomyoma which involved the
tomatos, malignant triton tumours and rare intracranial
rhabdoid tumours (Jakate et al., 1988; Kodet et al., 1991;
Burger and Scheithauer, 1994; Parham et al., 1994). Rhabdomyomas may be encountered as a component of
the malignant germ cell tumours of the pineal gland and
suprasellar region, particularly in immature teratomas.
Skeletal muscle differentiation can also rarely be seen in
gliomas and meningiomas (Ferracini et al., 1982; Burger and Scheithauer, 1994).
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